

## 整合素样金属蛋白酶与凝血酶 13 型抗体

产品货号： mlR5856

英文名称： ADAMTS13

中文名称： 整合素样金属蛋白酶与凝血酶 13 型抗体

别名： Cleaves the vWF multimers in plasma into smaller forms. Von Willebrand factor cleaving protease; A disintegrin and metalloproteinase with thrombospondin motifs 13; A disintegrin like and metalloprotease (reprolysin type) with thrombospondin type 1 motif 13; A disintegrin like and metalloprotease with thrombospondin type 1 motif 13; ADAM metalloproteinase with thrombospondin type 1 motif 13; ADAM TS 13; ADAM TS13; ADAM-TS 13; ADAM-TS13; ADAMTS 13; ADAMTS-13; ADAMTS13; ADAMTS13 protein; ATS13\_HUMAN; C9orf8; TTP; von Willebrand factor-cleaving protease; vWF cleaving protease; vWF CP; vWF-cleaving protease; vWF-CP; vWFPCP.

研究领域： 肿瘤 细胞生物 免疫学 信号转导 细胞粘附分子

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Pig, Cow, Horse,

**产品应用：** WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

**分子量：** 145kDa

**细胞定位：** 分泌型蛋白

**性状：** Lyophilized or Liquid

**浓度：** 1mg/ml

**免疫原：** KLH conjugated synthetic peptide derived from human ADAMTS13:401-500/1427

**亚型：** IgG

**纯化方法：** affinity purified by Protein A

**储存液：** 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

**保存条件：** Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

**PubMed :** PubMed

**产品介绍 :** This gene encodes a member of the ADAMTS (a disintegrin and metalloproteinase with thrombospondin motif) protein family. Members of the family share several distinct protein modules, including a propeptide region, a metalloproteinase domain, a disintegrin-like domain, and a thrombospondin type 1 (TS) motif. Individual members of this family differ in the number of C-terminal TS motifs, and some have unique C-terminal domains. The enzyme encoded by this gene is the von Willebrand Factor (vWF)-cleaving protease, which is responsible for cleaving at the site of Tyr842-Met843 of the vWF molecule. A deficiency of this enzyme is associated with thrombotic thrombocytopenic purpura. Alternative splicing of this gene generates multiple transcript variants encoding different isoforms. [provided by RefSeq, Nov 2008].

**Function:**

Cleaves the vWF multimers in plasma into smaller forms.

**Subcellular Location:**

Secreted. Note=Secretion enhanced by O-fucosylation of TSP type-1 repeats.

**Tissue Specificity:**

Plasma. Expressed primarily in liver.

**Post-translational modifications:**

Glycosylated. O-fucosylated by POFUT2 on a serine or a threonine residue found within the consensus sequence C1-X(2)-(S/T)-C2-G of the TSP type-1 repeat domains where C1 and C2 are the first and second cysteine residue of the repeat, respectively. Fucosylated repeats can then be further glycosylated by the addition of a beta-1,3-glucose residue by the glucosyltransferase, B3GALT1. Fucosylation mediates the efficient secretion of ADAMTS13. May also be C-glycosylated on tryptophan residues within the consensus sequence W-X-X-W of the TPRs, and also N-glycosylated. These other glycosylations can also facilitate secretion.

The precursor is processed by a furin endopeptidase which cleaves off the pro-domain.

**DISEASE:**

Defects in ADAMTS13 are the cause of thrombotic thrombocytopenic purpura congenital (TTP) [MIM:274150]; also known as Upshaw-Schulman syndrome (USS). A hematologic disease characterized by hemolytic anemia with fragmentation of erythrocytes, thrombocytopenia, diffuse and non-focal neurologic findings, decreased renal function and fever.

**Similarity:**

Contains 2 CUB domains.

Contains 1 disintegrin domain.

Contains 1 peptidase M12B domain.

Contains 8 TSP type-1 domains.

**SWISS:**

Q76LX8

**Gene ID:**

11093

**Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

细胞外基质蛋白

产品图片

